

CORTICOSTEROID THERAPY (PREDNISONE, PREDNISOLONE)

Information for people with Diamond Blackfan anemia, and their families.

Why would a person with Diamond Blackfan anemia take corticosteroids?

In Diamond Blackfan anemia (DBA), the bone marrow (the center of the bone where blood cells are made) does not make enough red blood cells. A special protein in red blood cells called hemoglobin carries oxygen to all of the organs in the body. When the number of red blood cells is low, the organs in the body do not get the oxygen they need.

Corticosteroids can help people with DBA make more red blood cells. This therapy is often one of the first types of treatments doctors recommend for people with DBA. Doctors do not know exactly why corticosteroids work, but about 80% of people with DBA who take corticosteroids produce more red blood cells.

What are corticosteroids?

Corticosteroids are powerful drugs used to treat many conditions. They are used in people with DBA to help the body make more red blood cells. Steroids are also used to stop allergic reactions and reduce swelling.

Is this the same type of drug that people take to build muscle?

No. The steroids that athletes and other people take to increase muscle are anabolic steroids. The steroids used to treat people with DBA are corticosteroids.

What is corticosteroid treatment like?

The goal of corticosteroid treatment is to take only what the person needs to keep the hemoglobin in the body at a healthy level (about 10 g/dL).

Doctors usually prescribe a certain dose of corticosteroids for several weeks. If the hemoglobin in the body increases, then the dose may be decreased slowly. If the number of red blood cells drops too far, the dose might be increased. When the number of red blood cells is at a healthy level, the dose will be kept the same or slowly reduced.

Over time, the body might not respond to the corticosteroids and they will not work as well. If this happens, the doctor might increase the dose or suggest a different type of treatment such as blood transfusions or stem cell transplantation.

When changing the dose of corticosteroids, the doctor will check the amount of hemoglobin in the body using a blood test called a CBC (complete blood count) and reticulocyte count (this tells if the body is trying to make its own red blood cells).

How long does it take for corticosteroids to work?

If corticosteroids are going to work, the number of red blood cells usually increases in 2 to 4 weeks.

Are there side effects of corticosteroid treatment?

Yes, corticosteroids have both short-term and long-term side effects. Low doses of corticosteroids may be helpful with minimal side effects, but use of high dose corticosteroids for a long time can cause serious side effects. People taking steroids must be watched carefully for side effects. Other treatments, such as red cell transfusions, should be considered if a patient requires high corticosteroid doses or if serious side effects develop.

For more information call:

1-877-DBA-NURSE





Possible side effects of short-term corticosteroid use:

- Upset stomach
- Increased blood sugar
- Increased hunger
- Behavior changes, trouble sleeping, irritability
- Increased risk of pneumonia, thrush (white coating in the mouth), and other infections
- Weight gain, salt and water retention
- High blood pressure
- Increased fat on the face (rounded face), upper back, and belly
- Stretch marks on the skin, acne, poor wound healing, increased and unusual hair growth

Possible side effects of long-term use (3 months or longer):

- All short-term side effects
- Poor growth in children (can be severe)
- Brittle bones (bones break easily, problems with hips and shoulder joints)
- Muscle weakness
- Diabetes
- Eye problems

What can you do to improve the success of corticosteroid treatment?

- Take steroids exactly as prescribed by your doctor.
- Take steroids in the morning (if prescribed once a day), and take them with food to prevent an upset stomach.
- If you miss a dose of medicine, take it as soon as possible. If it is almost time for the next dose, skip the missed dose and return to the regular schedule. Do not take a double dose or extra doses.
- Tell your doctor if you get sick. Your blood count might drop from the stress of being sick, and you might need extra doses of medicine or other treatment.
- Always check with your doctor before changing the dose or stopping this medicine. It can be dangerous to stop corticosteroids suddenly. Your doctor may want to slowly reduce the amount you're taking until you stop completely.
- Talk with your doctor about any concerns. Make a list of your questions. Do your research so you can help make decisions about your treatment.

Important things to remember:

- Call your doctor if you have a fever of 100.5°F more than once or if you have a fever of 101°F or higher even once. It could be a sign of an infection.
- Tell your doctor about any stomach pains.
- Ask your doctor about vaccinations. People taking a high dose of corticosteroids should not get certain vaccines, such as chicken pox (varicella) and measles, mumps, and rubella (MMR).
- Get a flu shot (not the nose spray vaccine) each year. Everyone living in your home should get a flu shot, too.
- Ask your doctor about taking an extra medicine (called PCP prophylaxis) to avoid getting a certain type of pneumonia.
- Get a vision check every year.
- Ask your doctor about getting a special x-ray, called a bone density scan, when you start taking corticosteroids. This will show the strength of your bones and help your doctor watch for bone loss.
- Ask your doctor about seeing a specialist (called an endocrinologist) to monitor growth and development while taking corticosteroids.
- Be aware that at times of hormone changes, such as puberty or pregnancy, the body's response to corticosteroids may change, and transfusion may be necessary.
- Ask for help. Support can come from friends, family members, doctors and nurses on your medical team, or a support group for people with DBA.

Resources

- For answers to your immediate medical questions, you can call the DBA nurse at 1-877-DBA-NURSE (1-877-322-6877).
- To connect with other people and families affected by DBA, visit the DBA Foundation at: <http://www.dbafoundation.org/home.html>
- For additional information and resources, visit the CDC at: <http://www.cdc.gov/ncbddd/hbd/dba/default.htm>